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Title: Primary Cutaneous Adenoid Cystic Carcinoma Of The Scalp Mimicking Cylindroma On Cytology And Squamous Cell Carcinoma Clinically: A Diagnostic Pitfall With Literature Review

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ABSTRACT

Background:
Primary cutaneous adenoid cystic carcinoma (PCACC) is an exceptionally rare neoplasm that poses significant diagnostic challenges. It may clinically mimic SCC due to ulceration, firm consistency, and occurrence in sun-exposed areas, cytological features on fine-needle aspiration cytology (FNAC) closely overlap with those of Cylindroma. Accurate diagnosis is imperative since PCACC and squamous cell carcinoma exhibit aggressive behavior despite an indolent initial course, demanding surgical excision and often adjuvant therapy.

Case Report:
We report the case of a 67year old female with a firm, raised scalp swelling with a clinical suspicion of squamous cell carcinoma (SCC). FNAC smears showed clusters of basaloid cells arranged around hyaline globules; the cytomorphology was reminiscent of Cylindroma. However, subsequent histopathological evaluation and immunohistochemistry confirmed the diagnosis of PCACC. Notably, thorough clinical workup excluded lesions in the salivary glands or breast.

Conclusion:
This report underscores the considerable overlap of clinical features between PCACC and SCC as well as FNAC features between Cylindroma and PCACC and highlights the necessity of integrating clinical, cytological, histopathological, and immunohistochemical data for accurate diagnosis. An extensive review of the literature demonstrates that misdiagnosis based on clinical findings and cytology alone may lead to improper management, given PCACC's potential for aggressive local invasion and distant metastasis.

Keywords :
Adenoid Cystic Carcinoma, Skin Neoplasms, Scalp Neoplasms, Cylindroma, Fine Needle Aspiration

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INTRODUCTION:

Primary cutaneous adenoid cystic carcinoma (PCACC) is rare tumor with an estimated incidence of just 0.23 cases per 1,000,000 people annually.[1] This rarity often leads to diagnostic challenges, especially when PCACC presents in atypical locations such as the scalp. Despite its low incidence, PCACC is characterized by infiltrative growth pattern, resulting in local recurrence and perineural invasion.[2] Conversely, cylindromas are benign, slow-growing scalp tumors. Both exhibit basaloid cells around hyaline material on FNAC. While histopathology and IHC remain gold standards, FNAC is often the first diagnostic tool in resource-limited settings. This underscores the need for heightened awareness of cytomorphological overlaps to prevent delayed intervention. This article explores the diagnostic complexities of PCACC, emphasizing cytological evaluation and the need for integrated diagnostic approaches in guiding appropriate treatment strategies and improving clinical outcomes.

CASE REPORT :

A 67-year-old female presented to the surgery outpatient department with a raised swelling on the scalp. She had a history of head trauma approximately 10 years prior, and the lesion had gradually progressed over time, accompanied by seropurulent discharge for one month, without any associated pain. On examination, there was a 4 x 4 cm globular, non-mobile, firm ulcerated swelling on the scalp. The lesion was clinically suspected to be squamous cell carcinoma (SCC) due to its ulcerated appearance and location on sun-exposed scalp skin (Figure 1).

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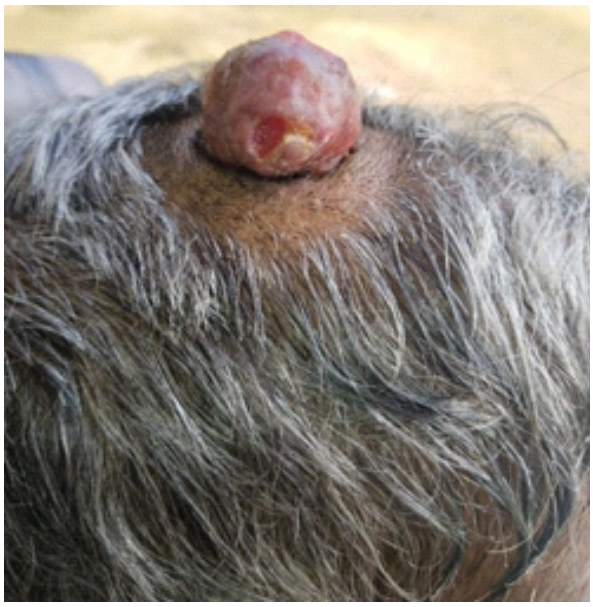

Figure. 1. Clinical presentation of the scalp lesion. A 4 × 4 cm globular, ulcerated, firm, and non-mobile swelling on the vertex of the scalp.

The FNAC smears stained with haematoxylin and eosin (H&E), Giemsa, and Papanicolaou (Pap), revealed high cellularity with multiple clusters of cells showing round hyperchromatic nuclei, inconspicuous nucleoli, and scant to moderate cytoplasm. The background displayed deposits of thick magenta-coloured basement membrane-like material and hyaline globules with palisading of tumor cells around them. Cribriform arrangement was also observed. PCACC and Cylindroma were considered as differential diagnoses due to overlapping cytological features (Figure 2).

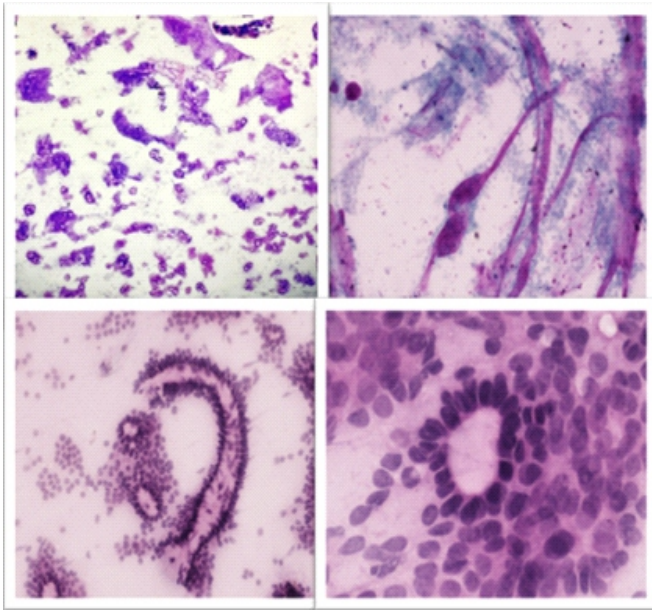

Figure 2:- Fine needle aspiration cytology findings (Clockwise from the left upper corner) low-power view (2×, H&E) showing hypercellular smears with cohesive clusters of tumor cells; thick basement membrane–like material with magenta-colored hyaline globules surrounded by tumor cells (20×, Giemsa); tumor cells with hyperchromatic nuclei and scant cytoplasm arranged around hyaline material with peripheral palisading (20×, H&E); high-power view (40×, H&E) highlighting basaloid tumor cells with marked nuclear hyperchromasia and minimal cytoplasm adherent to hyaline globules.

A wedge biopsy performed from the scalp swelling revealed a tissue with epidermis and dermis. The epidermis is intact. The dermis showed an unencapsulated and infiltrating tumor without any connection to the overlying epidermis in a desmoplastic stroma. The tumor is arranged predominantly in a cribriform pattern, with numerous punched-out spaces containing hyalinized material surrounded by basaloid cells. The tumor cells had round hyperchromatic nuclei, inconspicuous nucleoli, and a scant amount of cytoplasm. Mitosis was low (1-2/10 HPF). No perineural invasion noted even on multiple deeper sections examined. No keratin pearls or dyskeratotic cells were seen (Figure 3).

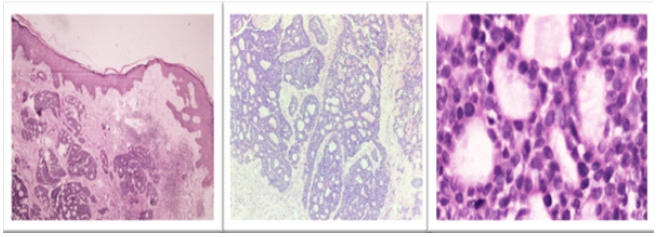

Figure 3: Histopathological features of primary cutaneous adenoid cystic carcinoma (Left to right): low-power view (4X, H&E) demonstrating an unencapsulated, infiltrative dermal tumor with no connection to the overlying epidermis; cribriform growth pattern with characteristic "punched-out" spaces containing hyalinized material (20×, H&E); high-power view (40X, H&E) showing basaloid tumor cells with scant cytoplasm and round hyperchromatic nuclei.

The immunohistochemical profile of the tumor demonstrated strong nuclear positivity for p63 (clone 4A4), and diffuse cytoplasmic positivity for pan-cytokeratin (AE1/AE3). In addition, focal faint cytoplasmic staining for CD117 (clone EP10) is also observed (Figure 4)

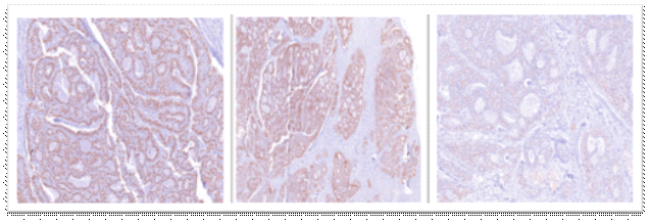

Figure 4:- Immunohistochemical features of primary cutaneous adenoid cystic carcinoma (Left to right): p63 immunostaining (20×) showing strong nuclear positivity in basaloid tumor cells, confirming myoepithelial differentiation; pan-cytokeratin immunostaining (10×) demonstrating difuse cytoplasmic positivity supporting epithelial origin; CD117 immunostaining (20×) revealing focal faint cytoplasmic positivity in tumor cells, aiding distinction from histologic mimics.

These findings collectively supported the diagnosis of PACC and aid in distinguishing it from other morphologically similar neoplasms. At 1.5-year follow-up, the patient remains disease-free without adjuvant therapy.

DISCUSSION:

PCACC is an exceptionally rare neoplasm that primarily affects the skin, often presenting as a slowly enlarging, firm, and occasionally painful nodule. PCACC is noted for its significant local invasiveness but generally has a more favourable outlook compared to salivary ACC. Although PCACC typically remains confined to the dermis, there are instances of distant metastases, usually to the lungs or regional lymph nodes, which can occur years after the initial diagnosis. This highlights the necessity for meticulous long-term surveillance.

The incidence of PCACC of is quite low primarily affecting middle-aged and elderly females. According to a population-based study in the United States, the incidence is around 0.23 per 1 million person-years. The scalp is the most common site of occurrence, accounting for about 41% of cases, with other less frequent sites including the chest, abdomen, back, eyelids, and perineum. While the Cylindromas constitutes 0.7% of all the adnexal tumors. This rarity and specific site predilection underscore the importance of accurate diagnosis.[3] Limited case reports, such as those by Yaranal PJ et al[4] and Vijayaraghavan A et al[5] illustrate the challenges in diagnosing PCACC. Our study, which involved scalp swelling, emphasizes the close resemblance between Cylindroma and adenoid cystic carcinoma, highlighting the need for careful differential diagnosis using FNAC. Other reports on scalp adenoid cystic carcinoma include studies by Temnithikul B et al[6], and Raja Tiwari et al[7] which were diagnosed through histopathological examination. Our case report is distinctive as it presents a diagnosis made using cytological analysis. The cytological, histopathological, and immunohistochemical features described in the present case and previously reported studies are summarized in Table 1.

Author	Age/ gender	Location of the tumor	Cytology	Histopathology	Immuno-histochemistry
Raja Tiwari et. Al[7]	65-years/ female	Right supra-orbital region	No FNAC reported	A poorly circumscribed infiltrative dermal tumor, lacking connection with the overlying skin. The tumor comprised basaloid and polygonal epithelial/ductal cells. Cystic spaces containing mucinous material and cellular debris noted. Prominent perineural invasion was observed.	Epithelial membrane antigen (EMA) and carcinoembryonic antigen (CEA) were positive.
Vijayaraghavan A et. Al[5]	48-years/ male	Forehead	Highly cellular smears with mainly cohesive clusters of cells surrounding eosinophilic hyaline globules. These cells were round, uniform, with minimal cytoplasm and tiny nucleoli, arranged in a small acinar pattern. The differential diagnosis included Adenoid Cystic Carcinoma (ACC) and the adenoid variant of Basal Cell	The tumor exhibited basaloid cell clusters in lace-like, cribriform, and glandular patterns. Basophilic mucinous and eosinophilic material was present within cystic spaces, and perineural infiltration was observed. Mitosis was noted at two per high power field. The differential diagnosis included cutaneous Adenoid Cystic Carcinoma (ACC) and the adenoid variant of Basal Cell Carcinoma (BCC).	Diffuse S100 positivity confirming the diagnosis of cutaneous Adenoid Cystic Carcinoma (ACC).

Yaranal et. Al[4]	55-years/Female	Medial malleolus of left leg	Tumor cells in clusters with round to oval nuclei and moderate eosinophilic cytoplasm Differential diagnoses included clear cell sarcoma, epithelioid synovial sarcoma, and amelanotic melanoma, with an excision biopsy recommended for further evaluation.	Sections revealed an infiltrating tumor with basaloid cells arranged in cords, nests, cribriform, and tubular patterns within the reticular dermis and subcutis, without epidermal involvement. Tumor cells were mildly pleomorphic, with moderate eosinophilic cytoplasm, vesicular nuclei, and occasional prominent nucleoli. Hyaline globules were present.	Epithelial membrane antigen (EMA) and vimentin highlighted the ductal component of the tumor, while the myoepithelial component was positive for S100 and cytokeratin. The tumor was negative for thyroid transcription factor-1 (TTF-1), carcinoembryonic antigen (CEA), and thyroglobulin.
Temnithikul B et. Al[6]	70-years/female	posterior aspect of scalp	No FNAC reported	The lesion was surgically excised, and histopathology revealed an adenoid cystic carcinoma (ACC) with perineural invasion.	positive for carcinoembryonic antigen (CEA)
Present case	67/female	Parietal convexity region on the scalp	highly cellular smears with basaloid clusters of cells around thick basement membrane-like material and hyaline globules.	Dermis showed an unencapsulated and infiltrating tumor without any connection to the overlying epidermis. The tumor is arranged predominantly in a cribriform pattern,	positive for p63, pan-cytokeratin and focal faint positivity for CD117

Table 1: Comparison of the Present Case Report with Previously Documented Cases of Primary cutaneous adenoid cystic carcinoma

Because of its quick turnaround and low invasiveness, FNAC is an essential diagnostic technique for scalp lesions, especially in settings with limited resources.[8] However, as demonstrated in this instance, there are substantial diagnostic difficulties due to the cytological overlap between cylindroma and PCACC. In our case, FNAC showed a blood-mixed, high-cellularity aspirate that initially suggested cylindroma due to the presence of basaloid cells grouped around hyaline globules, cribriform patterns, and peripheral palisading. Nevertheless, PCACC was confirmed by subsequent histopathology and immunohistochemistry (IHC), underscoring the limitations of FNAC. Based on this case and previous research, Table 2 contrasts the main cytological characteristics of PCACC and cylindroma.

Feature	PCACC	Cylindroma
Cellular Arrangement	3D clusters, cup-shaped structures, cribriform patterns	Tight sheets, vague acini, palisading arrangements
Nuclear Characteristics	Hyperchromatic, irregular membranes, nuclear molding	Round to oval, uniform, fine chromatin, inconspicuous nucleoli
Cytoplasm	Scant to moderate, occasional vacuolations	Scant, uniform
Basement Membrane	Thin, variable size, surrounding hyaline globules	Thick, attached to cellular clusters
Mucin Presence	Present (magenta-colored material on Giemsa)	Absent
Peripheral Palisading	Present, around hyaline globules	Present, around clusters
Background	Hyaline globules, basement membrane-like material	Hyaline globules, minimal background material
Mitotic Activity	Low to moderate (variable, often <2/10 HPF)	Rare to absent

Table 2: Cytological Features of Primary Cutaneous Adenoid Cystic Carcinoma (PCACC) vs. Cylindroma on FNAC.

Our smears (Figure 2) show that both entities exhibit peripheral palisading, which highlights their cytomorphological overlap. The lack of keratin pearls or atypical squamous differentiation made the diagnosis more difficult in this instance, ruling out SCC, but the hyaline globules and basaloid clustering looked like cylindroma. Histopathological confirmation was required to evaluate perineural or lymphovascular invasion, which is essential for PCACC diagnosis. Although precise misdiagnosis rates are still unknown, Amita et al[9] report that up to 30% of scalp FNACs may be mistakenly diagnosed as cylindroma because of shared features. Given its propensity for distant metastasis and local recurrence, this overlap can postpone PCACC diagnosis in resource-constrained settings where FNAC is frequently given precedence over biopsy . It is crucial to combine FNAC with early histopathological assessment and IHC (p63, CD117, Ki-67.

Accurate differentiation between PCACC and Cylindroma through cytological examination is essential due to their differing prognoses and therapeutic requirements. Both neoplasms share overlapping cytological features which can complicate cytological diagnosis. Cylindromas are often located in hair follicle-rich areas such as the scalp and can present as solitary or multiple lesions, sometimes resembling a "turban" due to their appearance as seen in our case. In contrast, PCACC has distinctive features including perineural invasion and a tendency for distant metastases.[10] Accurate differentiation is vital due to PCACC's aggressive nature and need for intensive treatment, including surgery and possibly adjuvant therapies. Misdiagnosis as Cylindroma, which is typically benign, can lead to inadequate treatment and worse outcomes due to delayed intervention and improper management strategies. Further studies are needed to estimate FNAC misdiagnosis prevalence. PCACC may clinically mimic SCC due to ulceration, firm consistency, and occurrence in sun-exposed areas. Notably, SCC typically presents with Keratin pearls and atypical keratinocytes on cytology which were absent in our case (Table 3).

Feature	PCACC	Cylindroma	SCC
Ulceration	Common	Rare	Common
Cellular arrangement	3D clusters, cup-shaped structures	Tight sheets, vague acini	Nests with keratinization
Basement membrane	Thin, variable	Thick, attached to clusters	Thin
Mucin	Present	Absent	Absent
IHC (Key markers)	CD117+, high Ki-67 index	CD117-, low Ki-67 index	CD117 not routinely tested, variable Ki-67 index.

Table-3: Key clinical, morphological and immunohistochemical features of PCACC vs Cylindroma vs SCC

Our case exemplifies how PCACC may masquerade as Cylindroma on FNAC due to shared features like hyaline globules and basaloid clustering. Notably, ACC often lacks the thickened basement membrane seen in Cylindroma, but this distinction is frequently obscured on smears. These findings highlight the necessity of integrating histopathology and IHC to confirm PCACC.

CONCLUSION:

This case highlights that PCACC, though rare, can clinically resemble SCC or other ulcerated malignancies. Although FNAC is a useful diagnostic tool, its ability to distinguish between primary cutaneous adenoid cystic carcinoma (PCACC) and Cylindroma is limited because of their substantial cytomorphological overlap. Early consideration of histopathological examination and immunohistochemistry, particularly markers like p63, CD117, and Ki-67, is highly advised in cases of scalp lesions with unclear cytologic features. By integrating these findings into clinical practice, healthcare providers can enhance diagnostic precision, optimize treatment strategies, and ensure effective patient care.

Conflict Of Interest: None.

Source of Funding : None

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Author Contribution:

RS, PN: Data acquisition, manuscript drafting. **AS,PR**: Data acquisition, manuscript review and editing. **LJ**: Reviewed the final version of the manuscript and approved it for publication.

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